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Ehrlichia Meningitis Mimicking Aneurysmal Subarachnoid Hemorrhage: A Case Study for Medical Decision-Making Heuristics

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Abstract

Thunderclap headache is a sudden and severe headache that can occur after an aneurysmal subarachnoid hemorrhage (SAH). Subarachnoid hemorrhage is a medical emergency that requires prompt attention and hospitalization. Patients with thunderclap headache often undergo a noncontrast head computed tomography (CT) scan to ascertain SAH bleeding and, if the scan is negative, then undergo a lumbar puncture to look for cerebrospinal fluid (CSF) red blood cells (RBCs), which would be consistent with an aneurysmal leak. If the initial CT is negative and CSF is positive for RBCs, patients are usually admitted to the hospital for evaluation of intracranial aneurysm. We encountered a patient with thunderclap headache whose initial head CT was negative for SAH and whose CSF tested positive for RBCs. The patient was referred to our center for evaluation and management of aneurysmal SAH. However, on careful review of the patient's medical history, serum laboratory values, and spinal fluid values, the patient was diagnosed with *Ehrlichia chaffeensis* meningitis. While *Ehrlichia* meningitis is rare, it is important to recognize the clinical clues that could help avoid formal cerebral angiography, a costly and potentially unnecessary procedure. We present how this case represented a cognitive framing bias and anchoring heuristic as well as steps that medical providers can use to prevent such cognitive errors in diagnosis.

Keywords

subarachnoid hemorrhage, cognitive heuristics, Erlichia

Introduction

Daniel Kahneman and Amos Tversky published a landmark article, "The Framing of Decisions and the Psychology of Choice,"[1] for which, along with their subsequent research, Kahneman was awarded a Nobel prize in Economics in 2002. This work is important to all physicians because it deals with errors in decision making based on ways information is provided and processed, including how physicians make medical decisions and how patients accept medical risks, such as surgery.[1,2]

Physicians often pride themselves on the ability to recognize medical history and physical examination patterns for accurate neurological diagnoses. However, data presentation can bias the physician based on certain cognitive preset values. The following case demonstrates what Kahneman describes as potential framing and anchoring errors in the clinical setting of a patient with thunderclap headache.[1,3]

Case Report

A 45-year-old right-handed man with a past medical history of asthma and previous cocaine and alcohol abuse presented to an outside hospital with thunderclap headache. The patient was seen in the outside hospital emergency department (ED) and underwent a noncontrast head computed tomography (CT) scan (Figure 1A), which was negative for SAH. Initial serum laboratory values (including urine drug screen) at this ED were reported to our neurosurgical team to be within normal limits, with the exception of platelets (86 000, normal = 150-450 000), alanine aminotransferase (229, normal = 7-55), aspartate aminotransferase (177, normal = 8-48), and alkaline phosphatase (141, normal = 45-115). A subsequent lumbar puncture demonstrated 1593 red blood cells (RBCs) in the cerebrospinal fluid (CSF), which could indicate possible SAH from a ruptured aneurysm, and 57 white blood cells (WBCs;

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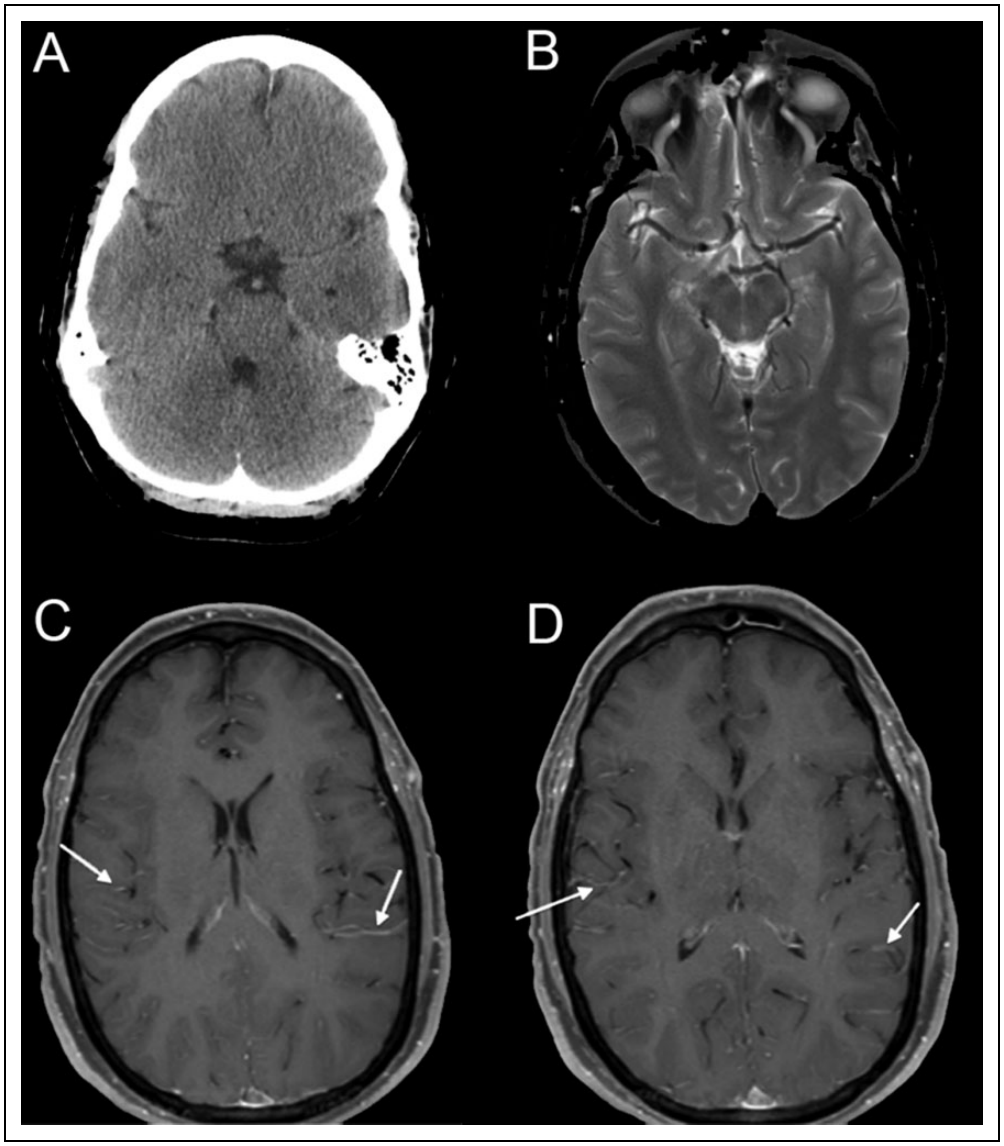

Figure 1. A, Noncontrast computed tomography (CT) scan of the head: negative for typical star-shaped SAH pattern. B, Magnetic resonance imaging (MRI) scan of the brain with and without contrast T2 at level of middle cerebral arteries. C and D, arrows pointing to leptomeningeal sulcal enhancement post-Gadolinium administration.

51% polynuclear WBC, 39% lymphocytes, and 10% mononuclear cells), 101 mg/dL CSF protein, and 69 mg/dL glucose (with a serum glucose of 138). Initial vital signs were Tmax 37.8°C, heart rate of 104 beats/min, 16 respirations/min, blood pressure of 137/64 mm Hg, and oxygen saturation of 100% on room air. Due to concern for aneurysmal SAH, a medical emergency, the ED physician called our institution at 1:35 AM to arrange a hospital-to-hospital transfer, secondary to possible need for aneurysm repair. He arrived to our institution at 4:35 AM under the care of the neurosurgical service and was seen by overnight Advanced Registered Nurse Practioner (ARNP). His vital signs upon admission were a temperature of 37.8°C, heart rate of 88 beats/min, blood pressure of 118/82 mm Hg, and respiratory rate of 16 breaths/min with 95% oxygen saturation on room air.

The initial differential diagnosis for his thunderclap headache included SAH or sentinel headache, reversible cerebral vasoconstriction syndrome (RCVS), or drug-induced RCVS, given his remote history of cocaine use, despite reporting no cocaine use for the last 10 years. Other causes of headache that were considered included cerebral venous thrombosis and an infectious cause such as viral meningitis or encephalitis. Due to concern regarding RBC predominance in CSF, despite normal noncontrast CT scan, the patient underwent CT angiography (CTA) and formal cerebral angiography. It was thought that his mild temperature elevation was secondary to central fever origin in the setting of SAH. The CTA was preformed within 1 hour of his arrival and formal cerebral angiogram performed within 2 hours of arrival. It was felt given the level of

acuity, RBC predominate CSF, and time-sensitive treatment options that, despite normal CTA, formal angiography was warranted by the neurosurgical team. This failed to show any intracranial aneurysm or vascular anomaly. There was no documentation regarding etiology or acknowledgment of his thrombocytopenia and transaminitis upon initial history and physical. The patient was transferred from the intensive care unit to the hospital neurology floor service for further evaluation.

On review of the clinical history by the neurology resident and attending neurologist, the patient endorsed acute onset of a nonpositional bilateral retro-orbital headache upon awakening, which was throbbing in nature and rated an 8/10 with maximal 10/10 at the time of onset accompanied by nausea, emesis, and photophobia 2 days prior to presentation to the outside ED. He denied any history of headaches in the past. He reported vacationing in Florida with his family who live in Georgia. His occupation was a pest and insect exterminator. The patient recently went camping in Georgia and had been bitten by "numerous insects." Questioning by the neurology resident disclosed a 1-week prodromal syndrome of diffuse arthralgia and fatigue prior to onset of headache. The neurology service evaluation took place 2 hours after formal cerebral angiogram was performed. Repeat examination by the neurology resident displayed a 24-hour maximum oral temperature of 39.5°C (103°F), mild tachycardia (94 beats/min), diaphoresis, and nuchal rigidity. Neurological examination was unremarkable for focal neurological deficits.

A revised differential diagnosis by the neurology service based on this additional history included potential central nervous system (CNS) infection, particularly herpes simplex virus (HSV) encephalitis, given the predominance of RBCs in his CSF, and rickettsia infection, given his travel and insect bite history. A repeat lumbar puncture was performed. The CSF continued to display RBC predominance (RBC = 9130, WBC = 113 with 9% lymphocytes, 86% neutrophil, and 5% mononuclear cells, glucose = 83 with serum glucose of 143, 81 mg/dL protein, negative Gram stain, culture showed no growth, and HSV polymerase chain reaction, Cryptococcal antigen, and histoplasmosis antibody were negative). The patient was started on broad-spectrum antibiotic and antiviral therapy. Infectious disease was consulted. Magnetic resonance imaging (MRI) was obtained to evaluate for possible abscess or herpes encephalitis, which displayed bilateral cerebral leptomeningeal sulcal enhancement (Figure 1 B-D). The CSF *E chaffeensis* PCR was positive. He was treated with 100 mg of doxycycline twice daily for 14 days to cover possible coinfection of *Borrelia burgdorferi*; however, Lyme serology was negative. The patient's symptoms normalized, and he had a complete recovery.

Discussion

This case illustrates the cognitive neuropsychology of medical decision making and evidence for framing bias. Framing bias occurred when the diagnosis of SAH was pushed to the top of the differential list. This can occur solely due to its common occurrence in the ED, despite historical clues that an alternative diagnosis might exist. Dr Daniel Kahneman's states in "Thinking Fast and Thinking Slow"[4] that neurocognitive decision making happens either through a fast, reflexive system called system 1 that occurs with minimal cognitive effort and represents the accumulation of accrued knowledge and experiences or through system 2 which is slower and more deliberate and requires more effort and concentration. System 2 is vulnerable to fatigue. By default, system 1 tends to predominate decisions in high-pressure situations. In this patient's case, the description of a sudden severe headache creates a framing bias using system 1 to find the simplest diagnosis compatible with CSF positive for blood. Because SAH is an emergency that requires prompt attention, it is understandable that system 1 would quickly offer this diagnosis. Considering the patient's low platelet count, history of potential insect bites would require conscious effort (system 2) and override a system 1 diagnosis of SAH. In a busy ED, this would require conscious effort to override the system 1 diagnosis.

Kahneman described the concept of framing effect or framing bias as when some clues are more heavily weighted than others in a decision-making process (Table 1**)**. This leads to a decision with the "easiest" conclusion. The ED physicians and neurologists weigh the diagnosis of SAH heavily so that it is not missed, which is crucial, given its potentially fatal outcomes. Therefore, the propensity to gravitate toward this diagnosis errs on the side of safety and availability. In this case, the prodromal symptoms, insect bite history, and laboratory findings should have been more heavily weighted. This is important because it could have led to less diagnostic testing and more cost-effective care.

Another important bias is anchoring, which pertains to the focus on the initial diagnosis and failing to adjust when new data is presented. In this case, SAH was anchored despite clues to the contrary (eg, thrombocytopenia and transaminitis on the initial hospital laboratory test results). These laboratory abnormalities were not mentioned in the initial admission history and physical, which may be due to the neurosurgery service being asked to accept the patient in order to perform a diagnostic and therapeutic intervention under the assumption of SAH diagnosis, which anchored them into their clinical role in the care of the patient.

Ehrlichia chaffeensis, the actual diagnosis, results in Human Monocytotropic Ehrlichiosis (HME) and was first described in 1987 by Maeda and collegues.[6] It occurs via the transmission vector *Amblyomma americanum*, commonly known as the lone star tick.[7] The majority of affected patients reside in rural areas with a median age of 44 years.[7] Infection causes a wide spectrum of illness ranging from asymptomatic to fatal. The major target cells for infection are monocytes and macrophages. Cases that have been fatal typically occur in immunocompromised patients or patients for whom treatment has been delayed after symptom onset.[7]

Central nervous system manifestations of HME occur in approximately 20% of patients.[8] Clinical CNS abnormalities range

Table 1. Cognitive Heuristics, Biases, Pitfalls With Examples, and Potential Interventions.[2,5]

Heuristic Type or Phenomenon	Pitfall	Examples	Interventions
Framing effects	Focusing on certain aspects of a case due to subtle wording	Focusing on "worst headache of my life" rather than accompanying symptoms	Reevaluate case from alternative perspective
Anchoring	Placing judgment on initial information and failing to adjust in the face of additional data	Our case focusing on RBC predominance only and not taking into account fever, nuchal rigidity, and red flags in history	Formulate second opinion through the use of others or by oneself. Evaluate probability data based on additional information
Availability Heuristic	Judging a case due to recent memory or impact of a previous case	Seeing a rare case and then subsequently testing for that case in all clinical settings	Verify with evidence-based medicine
Blind obedience	Following an opinion or course of action despite possible evidence to the contrary	Applying a protocol to every patient, despite clinical situation without regard to individual clinical scenario	Determine diagnostic test performance characteristics
Premature closure	Adopting a narrow-minded belief in a single idea	Focusing only on one diagnosis and not performing entire differential	Reevaluate case at a different time

from headache associated with photophobia, confusion, lethargy, hyperreflexia, cranial nerve palsy, seizures, ataxia, and nuchal rigidity.[9] Cerebral spinal fluid analysis in HME reported in the literature describes pleocytosis with lymphocytic predominance associated with increased concentration of protein.[10] One case of a pediatric patient reported a CSF analysis of 10 RBCs.[8] Risk factors for infection include residing in an endemic area such as Mississippi, Tennessee, Arkansas, and Georgia; an immunosuppressed state; and known exposure to a lone star tick. The greatest number of cases is reported between the months of May through August. However, cases have been reported year round.[11] Doxycycline is the recommended treatment and is generally well tolerated.

In conclusion, this case illustrates the importance of maintaining cognitive resilience against cognitive biases. Framing, as described by Vickery et al,[5(p426)] "refers to the error of initiating diagnostic reasoning by overvaluing an item of clinical formation that is presented early in the process." In this case, the patient's examination findings of nuchal rigidity, fever, and photophobia, paired with his risk factors for possible tick-borne illness, must alert medical providers to the possibility of meningitis as opposed to SAH. Elevated hepatic transaminases, leukopenia, and thrombocytopenia additionally support evidence of HME that must be noted. This case provides an example of the importance of maintaining an open cognitive process in diagnosing patients. Ultimately, medical providers and educators should be aware of these important heuristic principles due to their impact not only in medical decision making but also in reducing health care expenditures due to unnecessary testing.

Authors' Note

Dr Freeman serves as an associate editor for *The Neurohospitalist.*

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